



Standards of Care for Cystic Fibrosis in New Zealand

Standards of Care for Cystic Fibrosis in New Zealand Group
Medical Advisory Committee of Cystic Fibrosis Association of New Zealand
January 2010

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1. SUMMARY

Standards of care define the optimal service provision necessary to deliver the best outcomes for patients. This paper sets out the minimum standards of care for people with Cystic Fibrosis (PWCF) in New Zealand. The principal aim is to provide guidance to ensure that all PWCF have equity of access to high quality care regardless of where they live in New Zealand. This document will act as a template to provide robust guidance for the future configuration of CF services. In response to the geographic challenges facing the delivery of equitable health care in New Zealand, rather than focusing purely on structure and size of CF clinics the primary focus of this document is to outline minimum standards of care each individual with CF should have access to - irrespective of where they live.

The level of expertise required to both treat and delay the onset of complex multi-system complications in CF is best achieved through the involvement of a multi-disciplinary team of trained, experienced, specialist health professionals. Due to the limitation of staff and patient numbers in many New Zealand CF clinics, it is unlikely that all aspects of CF care will be able to be provided to all individuals locally. Individual DHBs will need to determine whether they are able to fulfill all the service provision requirements at a local level, or whether they will need to develop linkages or shared care arrangements with other centres with appropriate resources.

Where a DHB is able to fulfill all the requirements of service provision for a PWCF, they should be recognized as a Regional CF centre. This document includes recommended staffing ratios for patient numbers and core CF services expected at Regional CF Centres. The challenge for those involved in a model of shared care (PWCF and health professionals alike) is to ensure that an effective relationship between PWCF and their local CF team is seen as a priority and that any shared care that occurs with Regional CF Teams strengthens rather than weakens that relationship.

Once approved, this document will be published on the websites of clinical and lay organisations with an interest in CF including the Thoracic Society of Australia and New Zealand, Paediatric Society of New Zealand, Royal Australasian College of Physicians and Cystic Fibrosis New Zealand.

2. INTRODUCTION

The unique complexities of CF make the provision of care for PWCF especially challenging. It is a multisystem disease which, over time, presents different (often additional) health problems for those who have the illness. It is increasingly recognised that effective care of PWCF requires partnership between those with the disease, their families, local health care providers and multidisciplinary CF teams with greater expertise in more complex areas of CF care, or with access to more specialised investigations and treatments.

There are a number of basic standards for the provision of CF care which are generic and which have been espoused in the numerous recently published documents from many different countries. This document aims to provide a similar framework for services for PWCF living in New Zealand. In many other countries (US, UK, France, Germany, etc) the population is large (>10 million) and of sufficient density that many Specialist CF Centres have been established, each with staffing numbers to allow team members to maintain and develop their respective expertise. Typically, the Specialist CF Centre will provide care for PWCF living locally plus shared care and clinical support for PWCF who attend a smaller Local CF Clinic.

New Zealand, by contrast, has a population which is similar to the size of an average health region in the UK (just over 4 million) or the equivalent of one CF Specialist Centre in Australia. Just under half of people in New Zealand live in 6 urban areas of >100,000 people (Greater Auckland, Christchurch, Wellington, Hamilton, Dunedin and Tauranga). The other half of the population live outside of these areas. If we apply international recommendations, Auckland is the only New Zealand city with sufficient population to support a Specialist CF Centre. However, a single Specialist Centre is impractical, due to the geography of New Zealand; therefore a model of care appropriate for New Zealand needs to be developed.

Equity of access to good local and national expertise is a central tenet of health care in New Zealand, as is the need to provide health care in accordance with the principles of the Treaty of Waitangi. This document is intended to set a bench mark for the necessary composition of CF services, the minimum standards for routine evaluation, assessment and treatment of PWCF no matter where they live in New Zealand. It is proposed that these standards will be adopted by all services and centres caring for PWCF in New Zealand.

Thus, the specific objectives of this document are:

- To define standards for the provision of care for PWCF in NZ, which will work towards ensuring all people with CF have equitable access to effective and safe health care.
- To provide evidence-based health care principles for health care professionals, PWCF and their families and for health care administrators.
- To provide a basis for audit and quality assurance of health care delivery for PWCF in NZ.
- To inform health professionals, DHB managers and Ministry of Health strategists of the minimum requirements for services to care for PWCF.

This document focuses on systems for the delivery of care and no attempt has been made to address specific aspects of CF care as there are a number of clinical management guidelines already readily available to assist CF health care providers.

3. BACKGROUND

Cystic Fibrosis (CF) is the most common, recessively-inherited disease in New Zealand with a carrier rate of 1 in 25 and an incidence of 1 in 3,000 live births. This would suggest that there are approximately 20 children born each year with CF, and more than 500 people currently living in New Zealand with CF (Ministry of Health figures, unpublished).

CF is caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. This results in absence or dysfunction of the apical membrane CFTR protein which predominantly regulates chloride transport out of epithelial cells. The clinical consequence is a multi-system disease characterised by progressive pulmonary infection and damage, pancreatic dysfunction, liver disease that may progress to cirrhosis, infertility, gut motility problems, and elevated sweat electrolytes. Ninety percent of mortality and the majority of morbidity is related to lung disease resulting from bronchiectasis and chronic pulmonary infection¹. With the increase in survival, other complications such as CF-related diabetes and osteoporosis are commonly affecting the lives of people with CF².

New Zealand was the first country in the world to begin a neonatal screening programme for CF in 1981. For the last two decades neonatal "blood spot" testing has been a two stage screening procedure performed routinely on all newborns in New Zealand measuring blood immuno-reactive trypsin (IRT) followed by testing for the three commonest genetic mutations in those with high IRT levels. Carrier testing has been available to relatives/partners of those with CF and extended specific CF mutation analysis is increasingly available if required.

Outcomes for people with CF have improved dramatically over the past 50 years^{3,4}. Death from CF as a child is now an unusual occurrence and median survival is predicted to reach 40 years in the next decade with survival in males generally longer^{5,6}. Factors which have contributed to this increased survival include: improved management of the neonatal complication of meconium ileus, advances in pancreatic enzyme replacement therapy and nutritional support, development of improved techniques in airway clearance, the availability of more potent antibiotics and CF-specific centre-based care^{7,8}. There is evidence that PWCF who are under the care of larger specialist centres generally have better outcomes than those cared for solely in smaller centres⁹. This has resulted in an internationally agreed model of care where PWCF have some form of oversight and review regularly from a Specialist Centre CF team.

In 2002 the Australasian Cystic Fibrosis Data Registry recorded 358 people with CF in New Zealand. The average life expectancy in New Zealand was 29 years; compared with average life expectancies of 33-37 years for Australia, UK, & USA at that time^{10,11,12}. The reasons for this difference are not clear. It is hoped that through the application of the principles of care set out in this document that life expectancy for CF in New Zealand can be brought up to at least match international outcomes.

Most DHBs should have an age-specific team caring for their local population of PWCF in Local CF Clinics. However, in larger metropolitan centres consisting of multiple DHBs a single age specific clinic should provide CF care for that region rather than duplicating services for each DHB. All patients should be able to identify and have ready access to a Local CF Clinic. Success in driving up quality of care for PWCF in New Zealand hinges upon the establishment of a network of clinics which will ensure PWCF have ease of access to all aspects of multi-disciplinary CF care. The level of expertise required to both treat and delay the onset of complex multi-system complications in CF is best achieved through the involvement of a multi-disciplinary team of trained, experienced, specialist health professionals who see patients at a Regional CF Centre. Currently no such model exists in New Zealand. The aim of this document outlines what is needed to create a framework to deliver CF care that meets international standards for the delivery of CF care with ready access to the wide range of specialist support services.

A major strength in New Zealand is the extent of CF knowledge base that already exists amongst health professionals in clinics - both small and large. There are already well established links which will continue to develop between a number of local paediatric clinics and the Starship Hospital CF Team. Furthermore, the CF community is small enough to encourage close communication and cooperation. The primary focus of CF care for many will remain through local CF Clinics with every effort made to improve CF services throughout the country. However, where CF care needs can't be met in smaller centres, referral pathways and shared care arrangements need to be established with a Regional CF centre. Larger centres need to evolve to meet the Standards of Care set out in this document for a Regional Centre to establish this model of care for PWCF.

Funding streams to allow the travel of both CF teams and patients need to be established which will require inter-DHB cooperation, and is likely to form part of the evolving framework of Managed Clinical Network in partnership with Ministry of Health.

The challenge for those involved in a model of shared care, PWCF and health professionals alike is to ensure that an effective relationship between PWCF and their Local CF Team is seen as a priority and that any shared care that occurs with Regional CF Centres strengthens rather than weakens that relationship.

4. MINIMUM STANDARDS FOR CF CARE AVAILABLE TO PWCF

4.1. Core CF Care Requirements

Irrespective of the number of patients attending any given CF clinic the following should all be provided:

1. All PWCF must be under the care of a named consultant(s), whose clinical training and expertise is appropriate to the age of the patient. Collaborative working and discussion between adult and paediatric colleagues in any particular CF Clinic (or between Centres/Clinics) is expected. It is not considered acceptable for adult patients to continue under the care of Paediatric specialists. Similarly, children should not be under the sole care of an Adult Respiratory Specialist. In those areas where there is no identified Paediatrician or Adult Physician to care for PWCF, this should be rectified as a matter of the utmost priority by the local DHB.
2. All facilities should have, and adhere to, infection control principles; provide care appropriate to the age of the patient.
3. Accommodate unique aspects of care required by PWCF
 - a. They may have more frequent admissions to hospital
 - b. Admissions are often of longer duration than average
 - c. Recurrent and life-long access to appropriate facilities is required
 - d. Typically patients are more mobile during hospitalisation and require leave for work, recreation and/or study to maintain independence and quality of life when compared to other patients
 - e. Have specific infection control issues
 - f. Have specific dietary requirements
 - g. Admissions to hospital can be urgent but they are often planned

Multi-disciplinary care should be routinely provided for all PWCF. This document outlines the services and staff that need to be available to all people with Cystic Fibrosis in New Zealand. Whether this contact is achieved for patients locally or through shared care and Regional CF Centres will depend on circumstances such as expertise at their Local CF Clinic and the complexity of their disease. To ensure core services can be provided for all PWCF, Regional centres need to be established. Regional CF Centres must have complete CF Multidisciplinary Team staffing as listed below so that all PWCF have ready access to a clearly defined CF team with the resources to address all aspects of their care. For individuals living outside Regional CF Centres their team is likely to comprise of a combination of health professionals from both Local and Regional Clinics.

Personnel required for Regional CF Centre Multidisciplinary Teams

- Lead Consultant Physician or Paediatrician with experience in care of PWCF
- Nurse
- Physiotherapist
- Dietitian
- Social Worker
- Psychologist
- Pharmacist
- Microbiology Services

See below for the recommended roles and responsibilities of these core staff members.

4.2. Outpatient Clinics

The primary locus of CF care should be at a hospital with a CF clinic closest to where a PWCF lives. The CF clinic should be held in a designated area, which should be appointed to meet the needs of the clinic. PWCF should be seen in specific CF clinic sessions, with adequate staffing, room availability, and access to routine monitoring tests. Clinics should have established guidelines to prevent cross infection between PWCF, and the clinic environment should comply with these guidelines. CF clinics should be held in an environment suitable for the patient's age group. Mixing of adult and paediatric patients at the same clinic is not appropriate.

All PWCF should be able to be seen at least four times per year in a dedicated CF clinic. For PWCF routinely attending a Regional CF Centre, this will mean regular access to health professionals covering all aspects of CF care. For those attending smaller local CF Clinics the makeup of CF teams at these reviews will vary. To support patients and local CF Clinic health professionals outside Regional CF Centres there needs to be a well defined relationship with a Regional CF Centre tailored to individual clinic and patient needs to ensure all aspects care for these patients are met and that local health professionals are adequately supported.

To ensure this occurs, a Regional multidisciplinary team should review all PWCF at least annually and more frequently as determined by an individual patient's needs. This may involve patients traveling to Regional CF Centres and/or through combined clinics to be held locally and involving a visit from core members of the Regional Centre Multidisciplinary Team. As a minimum core members would include a CF clinician, nurse specialist, physiotherapist and dietician. Between visits Regional CF Centre support can be augmented through the use of the TelePaediatrics service video link system. CF clinic frequency and capacity should be sufficient to accommodate all planned routine reviews of patients, with some leeway for additional reviews if necessary. CF team members should have sufficient time within their job plans to attend all clinics.

The extent of Regional CF Clinic contact will be determined by an individual's "needs" as perceived by the patient, their family/whanau and members of the local CF team rather than geography.

Annual reviews

All PWCF will have an annual review performed, and it is expected that Regional CF Centre involvement would occur at this time. Wherever possible, the annual review should occur in a setting that enables the input from all members of a specialist CF team. Assessment will include a series of investigations performed and a summary of recommendations for future management be collated.

4.3. Inpatient care

A hospital providing inpatient care for PWCF must have:

- A clear infection control policy.
- Single rooms, mainly to prevent cross-infection, and preferably with private en suite toilet and bathroom.
- Hand washing facilities and alcohol-based hand rubs present in each patient cubicle.
- Patients with known B. cepacia complex or MRSA nursed under appropriate infection control precautions.
- Facilities for supervised physical exercise.
- Means of pastoral support and distraction in order to prevent the negative experiences associated with the isolation of curtailing inter-patient contact.

It is estimated that for a CF clinic of 50 patients under sole care, 3-5 beds will be needed on average. This will be influenced by the severity and complexity of patient illness and whether the hospital receives CF inpatient transfers from other CF Clinics.

Educational and physiotherapy facilities which accommodate cross infection control measures should be available as required.

Both inpatients and those receiving intravenous antibiotic therapy at home should be discussed at least once weekly in a multidisciplinary meeting involving members of the CF team and the medical and nursing team on the ward or in the community.

CF team staffing must be sufficient to allow for all inpatient PWCF to have their care reviewed as frequently as necessary during the admission.

4.4. Clinical Support Services for Care of PWCF

There are core services, which should be available to all PWCF at local CF Clinics. It is impractical to expect that all specialist services can be provided outside Regional CF Centres. Access to specialist CF services must be made available to all patients wherever they live in New Zealand, when they are needed. The requirements of a Regional CF Centre are extensive and as described in many other documents, include medical and paediatric specialty clinics and on site availability of opinion from sub-specialists experienced in the CF complications of their particular sub-specialities. These include endocrinology, gastroenterology / hepatology, infectious diseases, ENT surgery, general surgery, thoracic surgery, obstetrics and gynaecology, fertility services, psychiatry, clinical genetics, anaesthetics, ICU specialists, sleep and respiratory failure specialists, immunologist / rheumatologist, palliative care specialists, and lung and liver transplant services.

The core services listed below need to be available at Regional Centres. The degree to which Local CF Clinics provide these services will vary, largely influenced by clinic size and available personnel. When services cannot be provided locally, they need to be readily accessible for PWCF through Regional CF Centres.

Laboratory and Radiology

- Laboratory services with adequate expertise to support management of CF such as microbiology, genetics and sweat electrolytes.
- Specialist radiology support including multislice CT, vascular access support, ultrasound services, DEXA scans and interventional radiology such as bronchial arteriography and embolisation.
- Plain radiology; in paediatric services this includes radiologists experienced in “scoring” the plain chest radiograph using accepted scoring systems.
- Services for insertion of PICC lines under radiological guidance.
- Respiratory function laboratory including complex lung function testing (generally above the age of 7 years) and high altitude simulation testing.

Respiratory

- Specialists in Paediatric and Adult Respiratory Medicine with specific tertiary training in CF.
- Written protocols for assessment and management of all respiratory aspects of CF.
- CF-specific infection control protocols.
- Access to specialised thoracic radiology including CT scanning and interventional radiology.
- Specialised microbiological services with direct links with microbiology laboratory staff and specialist microbiology / infectious disease advice.
- Access to oxygen services.

- Access to sleep and NIV services: experience in initiating, and supporting non-invasive ventilation, both acute and longer term (as a bridge to transplant or for symptomatic palliative care).
- Access to paediatric and adult flexible bronchoscopy.
- Thoracic surgical service.
- Access to lung transplant service.

CF Related Diabetes

- Protocols for the assessment of glycaemic control, particularly at times of “stress”.
- Protocols for the management of CFRD based on the Australasian Nutritional Guidelines of 2006 on CF Australia website.
- Access to, and an established working relationship with, an endocrinologist and diabetes service with knowledge and the expertise in the management of CFRD.

Hepatology

- Regular measurement of liver function tests including liver synthetic function.
- Protocols for the investigation and management of CF related liver disease.
- Access to, an established working relationship with, a gastroenterologist with knowledge and expertise in the management of CF associated chronic liver disease, including management of the complications of hypersplenism and oesophageal varices.
- Access to the variety of imaging techniques required for the investigation of hepato-biliary disorders in CF.
- An established working relationship with a surgeon experienced in the management of hepato-biliary disease in CF.
- Access to a Liver transplantation service.

Gastroenterology

- Access to tests of pancreatic function including faecal elastase levels.
- Protocol for the prevention and non surgical management of DIOS.
- Access to tests to determine adequacy of pancreatic enzyme replacement therapy, as might be clinically indicated.
- Protocols for the use of PERT, based on the Australasian Nutritional Guidelines of 2006 on CF Australia website.
- Access to and an established working relationship with a surgeon with knowledge and expertise in the management of intra-abdominal complications of CF.
- Access to and an established working relationship with a gastroenterologist, with knowledge and experience of the intra-abdominal manifestations of CF.

CF related bone disease

- Protocol for the investigation and management of CF related bone disease.
- Access to DEXA scanning.
- Availability of serum vitamin D and vitamin K measurements.
- Established working relationship with an endocrinologist with expertise in the management of sex hormone deficiency.

ENT/ORL

- Regular clinical assessment of CF-associated upper respiratory tract disease.
- Working relationship with an ORL specialist with knowledge and expertise in the management of CF-related upper respiratory tract disease.
- Access to CT scanning of the upper respiratory tract.
- Access to audiology testing

Gynaecology

- Regular assessment to include inquiry about gynaecologic problems associated with CF.
- Assess to an established working relationship with, a gynaecologist with knowledge and expertise in the management of CF related gynaecologic problems.
- Protocols for the management of candidiasis, stress incontinence, menstrual irregularities, amenorrhoea and other gynaecologic problems associated with CF.

Fertility

- Patient education on topics of fertility.
- Patient advice on contraception and avoidance of STDs.
- Access to fertility testing facilities.
- Access to fertility services for both males and females.

Genetic Counseling

- Access to genetic counseling services.

Obstetrics

- Early discussion on sexuality, fertility, contraception, STDs, pregnancy.
- Assessment of intentions.
- Discussion of risk and other issues associated with pregnancy.
- Availability of genetic testing for partner.
- Review of treatment with respect to possibility of pregnancy.
- Early referral mechanism to obstetrics and obstetric medicine services.
- Close and on-going working relationship between the CF team, obstetric services and obstetric medicine services.
- Regular review of patient during pregnancy; this may include earlier than usual assessment of glycaemic control.
- Well formulated delivery plan.
- Careful attention to post-natal issues, including breast feeding, nutrition of the mother, drugs transferred in breast milk etc.

Surgical services

- Working relationship with general/paediatric surgeons experienced in the management of intra-abdominal complications of cystic fibrosis.
- Established working relationship with vascular surgeons experienced in the insertion of venous access devices.
- Established working relationship with thoracic surgeons with experience in the management of CF complications including pneumothorax and massive haemoptysis, and with a knowledge of the potential requirements with respect to transplantation.

- Established working relationship with ORL surgeons experienced in the management of CF related upper respiratory tract complications.

Transplant services

- Access to lung and liver transplant services.
- Working knowledge of the indications for referral for consideration for transplantation.

Echocardiography

- Skilled echocardiographers with particular experience in assessing right ventricular function and non-invasive assessment of pulmonary artery pressure.

Palliative Care

- All CF clinics should have support from established palliative care services.
- Access to expertise in the initiation of and support in use of non-invasive ventilation.

4.5. Staffing

Staffing of CF clinics will be largely influenced by clinic size. For larger centres with 50 or more patients, there are consistent international recommendations regarding appropriate clinic staffing levels for CF care^{13,14}. However, many local CF Clinics in NZ will have fewer than 10 patients and there is little guidance in the international literature on how such clinics should be staffed. As stated earlier, there must be an age-specific clinician identified for each individual with CF and as a minimum there should be a nurse, physiotherapist and dietician with protected time to deliver care to the local CF population (See Appendix 1 – for guidelines for FTE recommended/10patients). All team members will establish and maintain working clinical relationships with their colleagues around the country. They will be encouraged to be members of their professional group, and specifically a CF Special Interest Group (where such groups exist).

Specific roles may be provided by one or more persons, who each may have a special interest, specific training, accredited qualifications in CF, or a combination of these. It is important that all professionals providing CF care are aware of their areas of clinical strength and weakness within CF management and when appropriate, seek advice from more experienced colleagues.

In regions with fewer patients with Cystic Fibrosis, the CF specific roles will form only a small proportion of an individual's job description. In regions with more patients, some team members may have their full job involved with CF care. There is a need for all CF disciplines to share their learning and skills across regions, by visits, meetings, teleconferences, and common protocols. Ongoing training and education will be essential to maintain best standards of care.

Specialist / Consultant

Typically the Lead Professional for a CF Service will be a Consultant Physician or Paediatrician. The CF Centre Lead Professional will have overall responsibility for ensuring their CF Centre meets the standards set out in this document.

Patients attending a CF Centre should be under the primary care of a named consultant(s). The CF specialist(s) will:

- Be the named lead consultant for patients attending the clinic.
- Have protected time for CF care, in keeping with the size and needs of the local clinic population.
- Advocate on behalf of all patients.
- Advocate for the CF service as a whole.
- Keep up to date with routine CF practices.

- Be responsible for ensuring the local clinic has sufficient resource to maintain regular entry of data to the CFNZ PORT CF database.
- Contribute to managed clinical networking in accordance with local needs, and the basic principles of CF shared care models.
- Ensure that their CF services have adequate staffing for the core CF team roles, and alert their local management to shortfalls in staffing.
- Ensure that their junior medical staff obtain exposure to both inpatient and outpatient CF care, in keeping with overall training needs.
- Maintain working clinical relationships with colleagues at other centres/clinics providing CF care.
- Establish working clinical relationship with colleagues at their hospital, or at other centres/clinics for other (less commonplace) aspects of CF care (e.g. gastroenterology, endocrinology and diabetes, surgical interventions etc).
- Ensure that at times of planned absence a deputising colleague is nominated, and that they have sufficient CF experience to provide temporary cover. Where required, colleagues in other centres/clinics may be approached to support the local deputy as necessary.

Nurse

CF nurses are generally the focal point of CF clinics, and provide an invaluable point of contact for families.

The CF nurse(s) will:

- Have protected time for CF care - in keeping with the size and needs of the Local CF Clinic.
- Receive funding and time to attend professional courses and meetings.
- Be the first point of contact for families with CF related issues, referring appropriately to other members of the CF team if required.
- Coordinate care between patient and family/whanau, community services and hospital, both practically and through support and advice.
- Be involved in the overall management of the patients including advocacy, clinical management, support and advice.
- Participate in clinical decision-making and monitoring of patient care and offer day to day advice to ward staff for patients receiving inpatient therapy as well as to community-based health professionals who are caring for patients in an outpatient or community setting.
- Educate patients (and their families and carers/whanau) about CF, as appropriate for the PWCF's age.
- Support families/whanau when children attend schools, kindergarten etc.
- Play an important role educating other health care professionals.
- Have specific roles in certain critical times such as at diagnosis, transition from paediatric to adult care, changes in clinical status such as onset of Pseudomonas colonisation, the diagnosis of CF-related diabetes, discussion about lung transplantation and the onset of palliative care.

Physiotherapist

Physiotherapy is an essential part of routine CF care.

The CF Physiotherapist(s) will:

- Have protected time for CF care, in keeping with the size and needs of the Local CF clinic.
- Regularly assess each patient's respiratory status (including monitoring breathing rate and pattern, volume, colour and consistency of sputum expectorated and degree of dyspnoea at rest and during physical activity).
- Develop an individualised, acceptable, tolerable, and effective and efficient airway clearance regimen taking into account all relevant physical and psychosocial factors.

Modern physiotherapy in CF is primarily preventative and has to be incorporated into each patient's daily routine. Therefore physiotherapy should be carried out in a way that makes future cooperation likely and encourages adherence.

- Undertake regular appraisal of airway clearance therapy and adjunctive inhalation therapy.
- Provide ongoing education and training in the optimal use of individually selected airway clearance techniques and devices together with provision of advice regarding the cleaning and maintenance of equipment.
- Review and assess exercise capacity, tolerance and participation including the assessment of oxygen saturation at rest and during exercise.
- Encourage regular exercise and help provide an exercise programme.
- Assess postural alignment, chest mobility, muscle strength and endurance and injury prevention.
- Address musculoskeletal problems as they arise.
- Provide ongoing education of the patient/carers relating to appropriate use of physiotherapy including airway clearance therapy, adjunctive inhalation therapy
- Assist in the management of "stress incontinence".

Preferably the same physiotherapist should provide both inpatient and outpatient care to ensure continuity.

Dietitian

Optimal nutrition has been shown to be vital in maintaining wellbeing in CF.

The CF Dietitian(s) will:

- Have protected time for CF care - in keeping with the size and needs of the Local CF Clinic.
- Advise and educate patients and care givers about the principles of nutritional management in CF.
- Undertake individual assessments of nutritional losses, requirements and intake, and provide age-specific, individualised advice, supported by appropriate educational material on nutritional requirements at varying stages of health and disease and monitor pancreatic enzyme replacement therapy (PERT) and vitamin therapy.
- Communicate nutrition issues with the CF team.
- Be able to counsel families regarding feeding difficulties.
- Identify abnormal feeding patterns and behaviour and work with Psychiatric Liaison Services to help manage patients with eating disorders.
- Ensure they are familiar with differences in nutritional care between IDDM and CFRD.

Preferably the same dietitian should provide both inpatient and outpatient care to ensure continuity.

Social Worker

PWCF can often have challenging psycho-social needs, often specific to their condition.

The CF social worker will:

- Have protected time for CF care - in keeping with the size and needs of the Local CF Clinic.
- Provide a range of practical and emotional support to patients and their families.
- Undertake regular patient review to identify psychosocial issues such as emotional and relationship problems, financial stress, substance misuse problems, and educational and employment issues.
- Advocate on issues such as disability and income support, concession card eligibility, child disability allowance, and access to travel and accommodation subsidies associated with travel to Regional CF Centres.
- Provide emotional support to patients and families when health is deteriorating or at specific times and assist them to evaluate short and long term options eg both the patient and their partner reducing their working hours or stopping.

Preferably the same social worker should provide both inpatient and outpatient care to ensure continuity.

NB: The New Zealand CF Association Field Workers are also an important resource for meeting the psychosocial needs of PWCF and their families/whanau.

Psychologist

PWCF and their family/whanau are vulnerable to a range of psychological difficulties. The nature of the condition and its treatment, impact on childrens', adolescents' and adults' abilities to respond to ordinary developmental tasks and extraordinary life events. As the condition progresses, physical deterioration can further impact on psychological wellbeing and quality of life. Consequently, the CF clinical psychologist has an important role in:

- Early involvement in developmentally appropriate strengthening of coping mechanisms for infants, children and their families.
- Comprehensive assessment of, and intervention in, emotional, behavioural and psychological difficulties.
- Management strategies to improve adherence.
- Assessment of the patient's and family's psychological resources and support interventions before lung transplantation.

Preferably the same psychologist should provide both inpatient and outpatient care to ensure continuity.

Pharmacist

Each CF Centre should identify a member of the pharmacy staff who will develop an interest in CF-related pharmacology, and be a first point of contact for queries relating to CF treatments. They should:

- Monitor for potential drug interactions, allergic phenomena and adverse drug reactions.
- Provide education of patients and CF team members on pharmaceutical aspects of therapy.
- Monitor for adherence issues or difficulties for patients accessing specific therapies.
- Support the provision of home IV antibiotic therapy.

5. SHARED CARE

There is evidence that PWCF who are under the care of larger specialist centres generally have better outcomes than those cared for solely in smaller centres. This has resulted in an internationally-agreed model of care where PWCF have some form of oversight and review from a Specialist Centre CF team regularly.

Having defined the minimum requirements for all CF clinics, New Zealand's unique geographical and demographic constraints need to be acknowledged. We believe the ideal model of shared care is one where local CF Teams are supported by designated Regional CF Centres Existing CF teams around New Zealand will need to determine whether they have sufficient patient numbers and staffing resources to operate as a regional CF centre, or whether they will need to develop linkages with another DHB or hospital that fulfills the criteria for a Regional CF centre. Specific models of shared care (outreach clinics, or patients visiting the Regional CF Centre) will be best determined through close liaison between the local CF Team, the patient and their family/whanau and the Regional CF Centres. These may vary; even between patients attending the same centre/clinic and/or be different for the same patient over time. Notwithstanding the need for local variation, it is expected that all PWCF will receive direct multidisciplinary overview from the appropriate Regional CF Centre, on at least a 12 monthly basis, utilising the agreed shared model of care.

6. CLINICAL CARE, AUDIT, PERFORMANCE MONITORING AND PORT CF

Clinical care

Quality of life, life expectancy and other clinical outcomes have all been shown to improve when all clinics/centres strive to match the standards achieved in the best performing clinics/centres.

National protocols should be developed in the following areas, facilitating best practice in all clinics/centres:

- Management of the new CF patient.
- Detection and treatment of the initial *P. aeruginosa* infection.
- Detection and treatment of other bacterial infections – e.g. *B. cepacia complex*, *S. maltophilia*.
- Detection and treatment and management of atypical mycobacterium.
- Infection control.
- Dosing and administration of antibiotics (including monitoring antibiotic serum levels).
- Diagnosis and treatment of ABPA.
- Treatment of pneumothorax.
- Management of major haemoptysis including bronchial artery embolisation.
- Feeding by nasogastric tube or gastrostomy.
- Diagnosis and treatment of hyperglycaemia and CF related diabetes.
- Diagnosis and treatment bone disease.
- Diagnoses and management of CF related liver disease and portal hypertension.
- Palliative care for PWCF.
- Transition from paediatric/adolescent to adult services.

Audit

It is expected that all CF Clinics/Centres will regularly audit aspects of their performance against nationally agreed standards of clinical practice, and where appropriate implement changes to local services to improve areas identified as below the desired standard. Wherever possible these experiences will be shared with other clinics/centres around the country to streamline the overall process of sustaining the highest possible standards of care for PWCF in New Zealand.

Performance Monitoring and Port CF

In addition to local audit, it is also important for all clinics/centres to contribute to national performance monitoring, to allow comparison with other countries. This will provide an external validation to practices in New Zealand and will help identify aspects of CF which might require attention, but which may be generic within the New Zealand CF population. Anonymised results from Port CF will be published annually by CFNZ, to allow individual CF Clinics/Centres to evaluate their areas of strength and weakness against the median performances across New Zealand. Changes to clinical practice which that evolve from such reporting and reflection should be shared with other CF Clinics/Centres around the country. It will also be useful to compare the national statistics for New Zealand with those of other countries to identify weaknesses, strengths and/or gaps of the CF care which may need investigation or

addressing. The data could be used to advocate for improved services on behalf of the CF community.

7. TRANSITION TO ADULT SERVICES

All clinics will develop systems and policies to ensure that transfer of adolescent PWCF from Paediatric Services to Adult Services occurs in a timely, measured and sensitive way, addressing both the individual's concerns and those of the family/whanau. These policies and systems should not only address the process of transfer from Paediatric to Adult services, but also specific pastoral needs for teenage PWCF. This should include discussion about fertility and sexual health, tertiary education or career options, movement away from the parental home, and (where necessary) the likelihood of transplant and discussion of medium to long term prognosis.

Policies of transition and transfer should also include strategies to support parents of PWCF in gradually relinquishing their previous high level of involvement with and commitment to the day to day requirements of CF care, whilst maintaining appropriate levels of support for the adolescent PWCF.

NB: Specific details around planning for transfer are set out in "Transition to Adult Health Services for adolescents with chronic conditions" RACP Joint Adolescent Health Committee Guidelines and readers are referred there for more clinical detail (<http://www.racp.edu.au/page/health-policy-and-advocacy/paediatrics-and-child-health>).

8. TREATY OF WAITANGI AND TIKANGA IN PRACTICE

The principles of the Treaty of Waitangi, and the need for Tikanga in practice will already be set out by all DHB's. Adherence to these are an important part of CF care in New Zealand (and one which differs from all other countries). There are no specific areas of CF practice which create conflicts with these principles.

9. CF ASSOCIATION OF NEW ZEALAND

All PWCF will have access to the Cystic Fibrosis Association of New Zealand, which provides valuable support and advice and advocacy for individuals and the CF community. Local CF Clinics are very likely to already be actively engaged with CFNZ for the benefit of the local patients, but all CF Clinic/Centre teams are encouraged to develop strong working links to support the organisations activities – e.g. supporting educational sessions for parents/whanau, the production of CFNZ literature for PWCF, families/whanau, reports from professional meetings and conferences etc (www.cfnz.org.nz).

10. CONCLUSION

The level of expertise required to both treat and delay the onset of complex multi-system complications in CF is best achieved through the involvement of a multi-disciplinary team of trained, experienced, specialist health professionals. Due to the limitation of staff and patient numbers in many New Zealand CF clinics, not all aspects of CF care will be able to be provided to individuals locally. It is hoped that by setting out clear guidance on minimum standards of care for PWCF, both in terms of local resources and the development of clinical networks between Local CF Clinics and Regional CF Centres, future planning to meet the needs of PWCF across the country will be encouraged. It is recognised that the standards and thresholds set out in this document are not in place in all CF Clinics across New Zealand at present. It is also recognised that there has been no formal discussion with CF teams across the country to reach an agreement on where Regional CF Centres should be located. It is inevitable that there will be a need to increase staffing levels at Local CF clinics and Regional CF Clinics to accommodate increased demand for appropriate care and for shared care.

It is proposed that once this document has been adopted, a survey to identify the degree to which Local CF Clinics across the country achieve or do not achieve these requirements will be undertaken, and the results of this survey will then be used as a starting point to initiate discussions with the Ministry of Health.

Standards of Care for Cystic Fibrosis in New Zealand Group
Medical Advisory Committee of Cystic Fibrosis Association of New Zealand
August 2010

Appendix 1: Recommended Staff Numbers for Paediatric and Adult CF Centres

	per 10 pts (min requirement)	per 50 sole care patients	Per 50 shared care pts (i.e for professional who visits another centre)
Consultant 1/ Centre Lead	0.1 (0.2)	0.5	0.25
Consultant 2	0.1 up to 20 pts; (0.05 per 10 pts over 20)	0.3	0.15
Clinical fellow	0.1	0.5	0.25
RMO/Reg		0.3	0.15
CFNS	0.3 (0.3)	2	1
Physiotherapist	0.3 (0.3)	2	1
Dietitian	0.1 (0.2)	0.5	0.25
Social worker	0.1 (0.2)	0.75	0.35
Secretary	0.1 (0.2)	0.5	0.25
Data clerk	0.1 (0.2)	0.4	0.2
Pharmacist	0.1 (0.2)	0.5	0.25

These numbers reflect FTE. However it is preferable to have this FTE reside with as few members of staff as possible (i.e. 0.5 FTE = 1 physiotherapist working 5 sessions, and not 5 physiotherapists, each doing 1 session per week).

References

1. Ratjen F, Doring G. Cystic fibrosis. *Lancet*. 2003 Feb 22;361(9358):681-9.
2. Cystic Fibrosis Foundation. Patient Registry Annual Data Report 2003. Bethesda, Maryland
3. Corey M, McLaughlin FJ, Williams M, Levison H. A comparison of survival, growth, and pulmonary function in patients with cystic fibrosis in Boston and Toronto. *J Clin Epidemiol* 1988; 41: 583-591.
4. British Paediatric Association working party on cystic fibrosis. Cystic fibrosis in the United Kingdom 1977-85: an improving picture. *BMJ* 1988; 297: 1599-1602.
5. Dodge JA, Lewis PA. Cystic fibrosis is no longer an important cause of childhood death in the UK. *Arch Dis Child*. 2005 May;90(5):547.
6. Elborn JS, Shale DJ, Britton JR. Cystic fibrosis: current survival and population estimates to the year 2000. *Thorax*. 1991 Dec;46(12):881-5.
7. Mahadeva R, Webb K, Westerbeek RC, Carroll NR, Dodd ME, Bilton D, et al. Clinical outcome in relation to care in centres specialising in cystic fibrosis: cross sectional study. *BMJ* 1998 Jun 13;316(7147):1771-5.
8. Noone PG, Knowles MR. Standard therapy of cystic fibrosis lung disease. In: Yankaskas JR, Knowles MR, editors. *Cystic Fibrosis in Adults*: Lippincott-Raven Publishers; 1999. p. 145-173.
9. Johnson C, Butler SM, Konstan MW, Morgan W, Wohl MEB. Factors Influencing Outcomes in Cystic Fibrosis. *Chest*. 2003;123:20-27.
10. Cystic Fibrosis Foundation. Patient Registry Annual Data Report 2003. Bethesda, Maryland.
11. Australian Cystic Fibrosis Data Registry. 2003. Cystic Fibrosis Australia, North Ryde, Australia.
12. UK CF database, University of Dundee, 2007.
13. Cystic Fibrosis Standards of Care, Australia, 2008.
14. Kerem E, Conway S, Elborn S, Heijerman H; Consensus Committee. Standards of Care for patients with Cystic Fibrosis: a European Consensus. *J Cyst Fibr* 2005; 4: 7-16.

ENDORSEMENTS

This Standards of Care document is endorsed by the following professional bodies:

The Paediatric Society of New Zealand
The Australasian College of Physicians
The Thoracic Society of Australia and New Zealand

Authors and Consultants Interest Register

Name	Contact	Conflict	Assessment
Dr Richard Laing	Christchurch Hospital Private Bag 4710, Christchurch	Chair of CFANZ Clinical Advisory Panel Chair of Pharmac CF Advisory Panel	Close - Manageable
Dr Cass Byrnes	Starship Childrens Health Private Bag 92024 Auckland Mail Centre Auckland	Member of CFANZ Clinical Advisory Panel Member PHARMAC CF Advisory Panel Australasian Cystic Fibrosis Bronchoalveolar Lavage Study – previously received Australian National Medical & Health Research Council and NZ Health Research Council research grants for this project.	Close -Manageable
Dr Peter McIlroy	Nelson Hospital Private Bag 18, Nelson	Member of CFANZ Clinical Advisory Panel	Close -Manageable
Dr Jeff Brown	Mid Central Health P O Box 2056 Palmerston North	Member National Health Board	Remote -Manageable
Dr Sarah Currie	Hawkes Bay DHB Private Bag 9014 Hastings	none	n/a
Dr Julian Vyas	Starship Childrens Health Private Bag 92024 Auckland Mail Centre Auckland	Member of Medical Advisory Group, Newborn Screening Programme	Remote - Manageable
Dr John Kolbe	Auckland City Hospital Private Bag 92024 Auckland Mail Centre Auckland	Employee, University of Auckland “Contractor”, Auckland District Health Board; President, RACP (and associated committees etc) ; Past President, TSANZ Board Member, CICM; Member, CPMC Member (previous); CF sub-Committee, PHARMAC; Respiratory sub-Committee, PHARMAC; Contract Research, includes Pharmaxis Aradigm Vertex MAC, NZ LAM Foundation	Close - Manageable
Professor Keith Grimwood	Brisbane University	Life Member CFANZ Former President CFANZ Former member Clinical Advisory Panel CFANZ	Remote - Manageable
Mrs Nicky Churton	232 Williams Street Kaiapoi, Christchurch	Member CFANZ Parent of a person with cystic fibrosis	Nil, document altered completely since initial draft.
Mr Doug Churton	232 Williams Street Kaiapoi, Christchurch	Member CFANZ Parent of a person with cystic fibrosis	Nil, document altered completely since initial draft.
Vivienne Isles	Christchurch Hospital Private Bag 4710 Christchurch	Member of CFANZ Clinical Advisory Panel Received funding for conference leave/travel expenses from: Boehringer Ingelheim, GlaxoSmithKline, Cystic Fibrosis Association of New Zealand	Close - Manageable
Kate Russell	Cystic Fibrosis Association of NZ	CEO – Cystic Fibrosis Association of NZ	Close – Manageable
Rebecca Scoones	Starship Childrens’ Health Private Bag 92024 Auckland	Member of CFANZ Clinical Advisory Panel	Close – Manageable
Dr Mark O’Carroll	Auckland Hospital, Private Bag 92024, AMC, Auckland	Member of CFANZ Clinical Advisory Panel, Member TSANZ Branch Executive, Member CF Subcommittee PHARMAC, Contract Research, includes Pharmaxis, Aradigm, Vertex, Mpex and Boehringer Ingleheim	Close – Manageable